

A00 Thrombotic thrombocytopenic purpura in pregnancy: a diagnostic emergency

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Introduction: Thrombotic thrombocytopenic purpura (TTP) is a rare but important cause of thrombocytopenia and haemolytic anaemia in pregnancy. We describe a case of acute onset congenital TTP in pregnancy, successfully treated with plasma exchange (PEX) and delivered uneventfully by caesarean section under spinal anaesthesia.

Case report: A 40-year-old primigravida presented at 36 weeks of gestation with severe thrombocytopenia and haemolysis. She was asymptomatic other than some minor gum bleeding and intermittent epistaxis, and had no previous medical history. Initial full blood count revealed a platelet count of $12 \times 10^9/L$ with a haemoglobin of 112g/L, low haptoglobins and elevated lactate dehydrogenase. An initial diagnosis of TTP was made on the basis of these laboratory parameters and the clinical history, with alternative diagnoses of HELLP, preeclampsia and acute fatty liver of pregnancy considered unlikely. Two units of fresh frozen plasma (FFP) and methylprednisolone were administered immediately, and PEX commenced within 8 h. This continued for 3 days and resulted in a steady rise in her platelet count to $163 \times 10^9/L$. Induction of labour was attempted on day 4; however, this was unsuccessful. She proceeded to deliver a healthy female by uncomplicated category 3 caesarean section at 36+5 weeks of gestation, performed under spinal anaesthesia with no adverse sequelae. Her platelet count remained stable postoperatively, and she was discharged home on day 4 postpartum. A diagnosis of congenital TTP was confirmed several weeks later with a low ADAMTS-13 enzyme level (<1%) on admission blood testing and no detectable autoantibody.

Discussion: Thrombotic thrombocytopenic purpura is a rare, acute and potentially life-threatening disorder that typically affects women and can be precipitated by pregnancy, with an incidence of 1 in 200,000 maternities.¹ It comprises thrombocytopenia, haemolytic anaemia and microvascular thrombosis, and has an untreated mortality of 90%. Women presenting with a new diagnosis of TTP in pregnancy typically do so in the third trimester or postpartum period, and are more likely to have a congenital rather than acquired aetiology.² TTP should be suspected in a case of severe thrombocytopenia in pregnancy and the specialist advice of a haematologist sought as prompt treatment with plasma exchange is potentially life-saving. PEX comprises apheresis and replacement with donor FFP that is repeated daily and has reduced the mortality of TTP to 20%.¹ Importantly, platelet transfusion is contraindicated in TTP as it fuels the coagulopathy. With successful treatment and subsequent improvement in the platelet count, delivery by caesarean section can be safely performed under spinal anaesthesia.

References

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